

Cerebral Coenurosis

Report of a Case with Arteritis

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Summary. A case of the unusual clinical condition of cerebral coenurosis, the unique parasitic disease that is localized only in the CNS, is reported.

The patient had repeated attacks of transient hemiparesis due to intracranial arteritis which was demonstrated by several angiographic studies.

The CSF showed a discreet lymphocytosis and increased immunoglobulins. This inflammatory reaction which preceded the manifestations of the intracranial mass can be explained by the parasitic foreign bodies, the liberation of somatic and metabolic parasitic toxins and the immunological reaction of the organism. The arteritis of the basal intracranial vessels was probably secondary to the inflammatory reaction of the leptomeninges.

In presence of the rare association of meningovascular disease and an expanding intracranial lesion a parasitic condition due to a larval stage of Cestodes (Hydatidosis, Cysticercosis, Coenurosis) should be always considered

Key words: Coenurosis, cerebral - Arteritis with coenurosis - Cestodes, cerebral.

Zusammenfassung. Es wird ein ungewöhnliches zerebrales Krankheitsbild, welches durch eine Cönurose hervorgerufen war, beschrieben. Bei der seltenen Kombination einer meningovaskulären Läsion mit einer intrakraniellen Raumforderung soll immer die Möglichkeit einer Zestodenerkrankung (Echinokokkose, Zystizerkose, Cönurose) erwogen werden. Von diesen drei Krankheitsformen befällt lediglich die Cönurose isoliert das Gehirn.

Die wiederholt angiographisch festgestellte Arteriitis der großen Hauptstämme äußerte sich klinisch durch wiederholte Halbseitenlähmungen.

Der lumbale Liquor zeigte jeweils eine geringe Lymphozytose und eine Vermehrung der gamma-Globuline und der Immunglobuline. Diese entzünd-

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liche Reaktion wird durch die Anwesenheit der Parasiten als Fremdkörper, durch die Freisetzung von Toxinen und durch die immunologische Antwort des Wirtes hervorgerufen.

Die Arteriitis selbst ist durch das Übergreifen der chronischen Leptomeningitis auf die Gefäßwand bedingt.

Cerebral coenurosis is a rare parasitic disease, specific for the CNS. Since the first description by Brumpt [4] in 1913, there have been 27 reports of intracranial manifestations in the literature. The case presented here was characterized by the association of meningeal, arterial and parenchymatous lesions.

Case Report

An Italian housewife who lived in a small town of French Switzerland for many years was in excellent health. She did not have any contact with domestic animals or keep dogs and had not traveled abroad for several years.

In 1974 at the age of 37, she began to suffer from left frontal headache. Shortly thereafter, she had four transient episodes of left hemiparesis.

First Admission to Hospital in May 1974. The general medical examination was normal.

Neurological examination disclosed slight right papilledema and a left central facial paresis. Laboratory Findings. Sedimentation rate normal, no anemia, 7150 leucocytes with 1.5% eosinophils. The plasma chemistry and urinary examinations were normal.

EEG showed an interhemispheric assymetry with frequent theta and delta waves of low voltage in the right precentral and parietotemporal area.

CSF. 24/3 cells with 95% lymphocytes, some eosinophiles and lymphoplasmocytes. Protein 440 mg/l. Glucose 55 mg%. The Lange gold curve showed a left deviation. Reactions for syphilis were negatives.

Skull X rays were normal.

Gammaencephalography with 99mTc showed a hypoperfusion of the right MCA territory during the early stage of the examination. The late stage was normal.

Right carotid angiogram (Fig. 1) showed a progressive stenosis of the distal portion of carotid siphon with a severe stenosis of the MCA. The A1 and the M1 segments were very thin. There was a generalized hypoperfusion of the MCA territory.

Left carotid angiogram showed an irregular narrowing of the M1 segment just before the M1—M2 angle. The A2 segment was also slightly narrowed.

The diagnosis of transient ischemic attacks in the MCA territory was made.

The patient was treated for several months with anticoagulants and the focal attacks disappeared.

In March 1976 the violent and persisting left frontotemporal headaches reappeared. Thereafter psychological changes occurred, she became drows, concentration was poor. She had olfactory hallucinations. In May 1976, she developed left ptosis with diplopia. These symptoms led to a second admission to hospital.

Second Admission May-July 1976. Neuropsychological examination disclosed an impairment of a verbal memory and a psychomotor dullness without aphasia or apraxia symptoms.

Neurological examination revealed a partial left oculomotor paresis with slight mydriasis and slow photomotor reaction. Except for a slight left central facial paresis, which was present in 1974, the cranial nerves were normal.

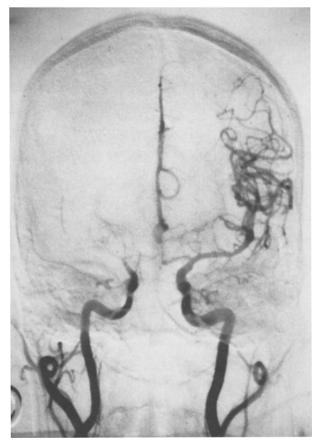


Fig. 1. Bilateral carotid angiography, May 1974. Stenosis of right carotid siphon with narrowing of anterior and middle cerebral arteries. Moderate stenosis of left middle cerebral artery

There was a slight weakness of the right arm and the deep reflexes were slightly more pronounced on the left. Coordination and the sensibility were normal.

Additional Findings: sedimentation rate normal, 6400 leucocytes with 2% eosinophils. Routine blood chemistry was normal, including electrophoresis and immunoelectrophoresis. The rheumatoid factors, Latex test, C reactives proteins, LE cells and antinuclear antibodies were negative as were the Mantoux and the Kweim tests.

The indirect immunofluorescence for Cysticerosis and Toxocariosis was negative. The passive hemagglutination for hydatidosis was slightly positive (1/180) as well as indirect immunofluorescence (1/10).

The skull X-rays were normal, as was the Doppler echography. *The EEG* showed a diffuse abnormality with polymorphous theta and delta waves in both parietotemporal areas. *Gammaencephalography* with ^{99m}Tc showed hypoperfusion in the left MCA territory during the early stage. In the late stage there was hyperfixation of the isotope in the left temporal area.

Right carotid angiogram showed a high grade of stenosis shortly after the beginning og M1. The parietal branches were well defined, while the small medial branches of MCA were not.

Left carotid angiogram (Fig. 2) showed a pronounced elevation of the sylvian group while the ACA was shifted to the right. There was a 60% stenosis in the middle part of M1 with a relative

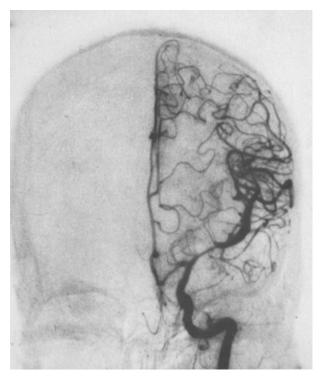


Fig. 2. Left carotid angiography, June 1976. Elevation of carotid siphon and group of the middle cerebral artery. Stenosis of middle cerebral artery

widening of the distal portion of the M1 and the M1-M2 angle. There is a "vessel free space" in the centrotemporal area.

Computed Tomography (CT scan). In the anterior part of the internal capsule as well as in the nucleus caudatus on left side there was a large cyst and bilateral hydrocephalus due to obstruction of the intraventricular foramina.

All cerebral structures were shifted to the right by the expanding cystic lesion.

CSF. 23/3 cells with 55% lymphocytes and 45% monocytes, without eosinophils. Protein 290 mg/l. Gammaglobulins were elevated to 47.8 mg/l (normal: 7—24 mg/l). Electrophoresis on agar gel showed an oligoclonal distribution in gamma 1-2-3 zones. Immunodiffusion showed an increase of gammaglobulins with 7.2 mg% of IgG (normal 0.8—2.5) and 1.1 mg% of IgM (normally not present in CSF).

The indirect immunofluorescence and the passive hemagglutination test for ecchinococcus and cysticercus were negative as well as the direct microscopic research for scolex or hooks.

On June 24, 1976 a left temporal craniotomy was performed. One cm under the second temporal convolution, there was a cystic cavity 1 cm in diameter. Under this cyst there was fibrous tissue which partly enveloped the sylvian group and extended to the sphenoid bone without adhesions to the dura. Laterally the fibrous tissue extended to the middle of the temporal lobe. At this point another cyst the of size of a cherry was punctured. The cystic fluid was clear and after the puncture the cyst wall could be completely removed. Exploration of the large cyst corresponding to the tomographic finding was not possible for technical reasons.

The histology of the first fragment, called "tumor-like tissue" showed a granular tissue rich in cells, with small necrotic areas surrounded by lymphoplasmocytes and eosinophils. No giant cells were seen. Vascularization of the granular tissue was very rich, consisting of capillaries and

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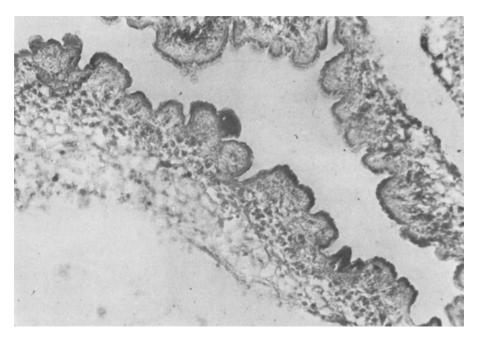


Fig. 3. Histology of wall of the parasitic cyst. H.-E., 160× numerous papillary villi of the inside of the cyst wall; every villi will later transform into a protoscolex

arterioles with partly thickened walls. Surrounding cerebral tissue there was a significant gliosis consisting mainly of protoplasmic astrocytes with spongiosis and microcalcifications.

Histology of the second fragment, called "cyst-like tissue" showed an inner amorphous layer, a middle small cell thiner layer and a loose fibrous outer layer. The inner layer presents characteristic villi typical of coenurosis (Fig. 3). The immediate postoperative course was without complications, but the patient had severe right hemiparesis and motor aphasia. Ten months later there was slight improvement of the neurological impairment.

Discussion

Coenurosis is caused by the larval stage of the Taenia multiceps multiceps. This worm, like the Taenia solium, the larvae of which provoke cysticercosis and like Taenia echinococcus the larvae of which are responsible for hydatidosis, belongs to the group of Cestodes which are Plathelmints [6, 11, 12, 15]. The definite host of adult worms is mainly the dog.

Taenia multiceps multiceps is characterized by a protoscolex with 4 suction cups and a weak rostrum armed with 23—32 hooks of different sizes. The adult worm has a length of 40—60 cm. The ovophoras are eliminated in dogs feces and can develop, if they are ingested by an intermediary host, like some herbivores, mainly the sheep. Human beings can be an intermediary host.

The ingested embryophores, 20—30 microns in size, spread through the whole body by the blood way. The larval development can take place only in the CNS and the eye [4, 7]. In these specific tissues the larva can develop to a well defined anatomical and histological form of a coenurus.

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During the proliferation stage, when the protoscolex are not yet formed, the coenurus wall is characterized by a typical papillary villiform pattern of the inner layer of the cyst. Every small villus will grow to an independent protoscolex which is a miniaturized adult worm which can grow only in the digestive tract of a dog. This microscopic aspect of the coenurus is typical and different from the cysts of other cestodes, notably cysticercus and hydatid.

From clinical point of view it is known that the coenurus is localized solely in the CNS and the eye, while the cysticercus and the hydatid invade several organs [6, 12, 15].

There is no specific biological test for coenurosis. Occasionally, there occur immunological cross reactions between the 3 representatives of cestodes, as shown by our patient, where the passive hemagglutination and the indirect immunofluorescence for ecchinococcus were slightly positive in the serum.

The pure ocular form of coenurosis (4 observations) and the localization in the spinal cord (a sole observation) are extremely rare. The intracranial form (28 observations) is the most usual one. We can differentiate in these cases between a meningeal, parenchymatous, vascular (seldom) and a combined form [1, 8]. The isolated menigeal attempt has been found only in children [8]. The cysts develop supra- as well as infratentorialy. They are frequently located at the base of the brain building arachnoid cysts [2, 3, 9]. Less frequently, the cysts are located inside the ventricles or in the brain parenchyma. The neurological symptoms correlate with the specific localization. Focal epileptic seizures were reported six times.

A further clinical manifestation is the occlusive hydrocephalus caused by the chronic menigenal inflammation, with the consequent obstruction of the arachnoid space or by an intraventricular cyst.

It should be noted that in the subarachnoid space there are usually several cysts, while in the cerebral parenchyma there is a solitary cyst. The presence of parasites in the subarachnoid space or in the brain tissue produces a local as well as a general inflammatory reaction which starts immediately after the parasitic invasion. It is caused directly by the presence of parasites as foreign bodies, but also by the liberation of somatic and metabolic toxins and by the immunological reactions of the infected organism.

The first manifestation of the chronic inflammatory reaction in CSF is a chronic basal meningitis [2, 3, 9] that manifests itself not only by menigeal signs, but particularly by the imflammatory modification of the CSF. These findings consist in slight pleocytosis with lymphocytes and plasmocytes. Eosinophils are occasionally observed [10, 14]. Proteins and glucose can be normal.

Our patient showed a considerable rise in her CSF gammaglobulins, mainly the IgG and IgM which underlines the importance of the immunological host reaction.

The illness of our patient was typical. She was in a good health and then had moderate neurological signs for two years before the appearance of specific symptoms of the intracranial space occupying lesion.

In addition to the chronic manifestation, the intermittent and remittent course should be mentioned. Finally, the combination of menigeal, vascular and parenchymatous signs seem to be very characteristic for a cestode disease. In our Cerebral Coenurosis 271

case, the meningeal (headache) and vascular symptoms (transient motor hemiparesis) started two years before the manifestation of the cysts.

This sequence of events was demonstrated by the repeated angiographic examinations.

Changes of the small vessels are frequent with diseases due to cestodes. A thickening of the adventitial layer with lymphoplasmocytic infiltration and atrophy of the muscular layer with intimal proliferation are frequently found. These modifications are the result of toxic damage of the vessel wall produced by necrosis of the neighboring parasites [17]. Although in our patient the large vessels (carotid artery, anterior and middle cerebral artery) were first and most severely involved we are of the opinion that this proximal intracranial arteriopathy is the consequence of the leptomeningeal inflammation similar to the pathogenesis in the case of the meningovascular syphilis.

This proximal arterial lesion is quite a rarity [17].

There is no specific treatment for coenurosis. Neurosurgery is successful only in cases with small parenchymatous or intraventricular solitary cysts. The intervention in the presence of multiple subarachnoid cysts is not always indicated and it can usually only relieve the effect of increased intracranial pressure. The prognosis is particularly bad if the cyst is accidentally ruptured during the intervention and the scolex are disseminated through the CNS. It is best to perform the operation in two stages. At the first intervention an alcohol or formol solution should be instilled into the cystic cavity. At second intervention, performed 24—48 h later the cyst can be safely extracted without danger of disseminating living scolex. In practice more important than these almost heroic therapeutic procedures, there should be prophylactic approach to parasitic diseases. In fact the infected dogs, which are the transmittors of the scolex, should be rapidly identified.

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